

REPORT OF A CASE OF MUSCULAR ATROPHY,
WITH PATHOLOGICAL FINDINGS IN SPI-
NAL CORD.¹

By W. H. RILEY, M.D.,

Battle Creek, Michigan.

THE patient was an American by birth, traveling salesman by occupation ; age forty-four ; came under my care for treatment of weakness in lower limbs, and gave the following history :—

His father had epilepsy when a boy, and again in old age. There was no history of rheumatism, syphilis, gout, or other chronic disease in the case. Habits good. He had been exposed some to the inclemency of the weather in his occupation as a traveling salesman for some years. His present trouble began about nine months ago, at which time he noticed a tendency on the part of the toes to drop and drag on the ground when walking. This was first noticed in the left leg, and in a few weeks in the right. The weakness of the muscles on the anterior of the leg increased, and others became affected, until at the time of my examination the patient was obliged to use crutches to get about. He also complained of a twitching of the muscles of the legs and trunk. He had never had any pain since this trouble began, and had not discovered any loss of sensation in the limb or any other part of the body. He tired easily on the slightest exertion, and had lost several pounds in weight since the beginning of the present trouble. Examination revealed a decided weakness in the flexors of the ankles and extensors of the toes. This was more marked in the left than in the right limb. There was also weakness of all the other muscles of the lower limbs. The muscles of the trunk were affected to that extent that it was with difficulty that the patient could rise from a horizontal to an upright position. At the time of examination there was no apparent weakness or

¹ Presented at the meeting of the American Neurological Association, New York, June 22, 23, 24, 1892.

wasting of the muscles of the upper extremity. There was a distinct fibrillary twitching in the muscles of the legs and trunk. This was constantly present. The knee-jerk was absent in both limbs, and there was no reflex action from stimulation of the skin in any part of the legs. Electrical examination showed the faradic irritability of the muscles of the anterior of both legs diminished, particularly those of the left side. A. Cl. C. equals C. Cl. C., in the peroneal muscles of the left leg. The sphincters were not involved. Sensibility to touch, pain, and temperature were normal, and there was no affection of the organs of special sense—hearing, vision, taste, and smell were all normal. Physical examinations of the heart, lungs, and abdominal viscera revealed nothing abnormal. Digestion, with the exception of constipated bowels, was fairly good. Diagnosis of progressive muscular atrophy was made, with an unfavorable prognosis.

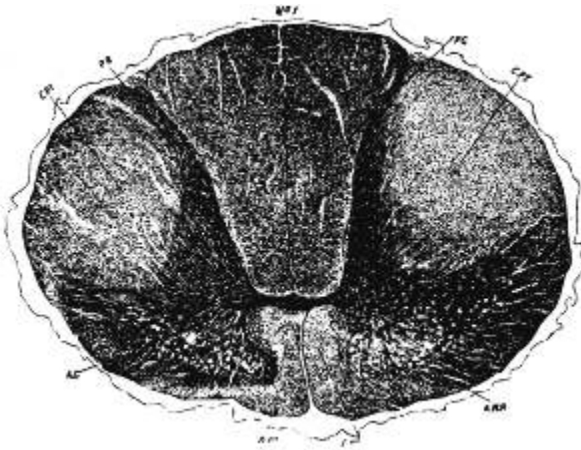
The patient was placed under treatment, consisting of massage, faradization, and galvanization of the muscles, together with the continued application of heat to the spine for one hour daily, and a galvanic current of forty milliamperes being passed directly through the spinal cord three times a week. Plenty of fresh air, sunshine, a nutritious diet, and rest in bed, with a little exercise, were prescribed. Under this treatment the patient got along comfortably for about two months, but made no improvement. The next month he seemed to grow gradually weaker, and at this time there was noticed a weakness and wasting of the thenar muscles of the right hand, which was soon followed by weakness and wasting of the corresponding muscles of the left hand. At the end of three months the patient was obliged to take his bed. There was a gradual progress of the disease. The wasting and weakness of the muscles first affected became extreme in degree, while new muscles were constantly being invaded by the morbid process. The diaphragm was finally paralyzed, and respiration was carried on by the thoracic and intercostals, and by the so-called muscles of "extraordinary inspiration." At this stage of the disease the patient was extremely emaciated and entirely helpless. During the last three or four weeks of the

disease the temperature varied from 98° to 100.8° F., pulse 120 and very weak. The amount of urea excreted daily gradually diminished during the progress of the disease, and a few days before death was about half in quantity to that excreted when the patient was first examined. Oxalates and phosphates of lime were present, and gradually increased in quantity. Fibrillary contractions in the muscles was a prominent symptom throughout the disease, nearly all the muscles being in a constant state of agitation and activity. The thoracic muscles, the *scaleni*, and the *sterno-cleido-mastoid* muscles of the neck, upon which the respiratory movements were entirely dependent at this stage of the disease (the diaphragm being entirely paralyzed), grew weaker and weaker, and the respiratory movements were only carried on with the greatest difficulty. The patient finally died on February 22d, about one year from the onset of the disease, the immediate cause of death being paralysis of the muscles of respiration.

Post-mortem examination was made. The muscles of the body were found shrunken and atrophied. There were streaks of light tissue through them, indicating a degeneration of the muscular fibre itself. There seemed to be no change in the internal organs, either abdominal or thoracic, except a general atonic condition of the tissues. The brain and spinal cord were examined. The brain appeared to be healthy. Microscopical examinations were made of the cord in the lumbar, dorsal, and cervical regions. These sections were hardened, and stained with Weigert's stain. The anterior cornua of the gray matter did not take the stain as readily as the posterior. There was a granular, broken-down appearance in the anterior horn, with the total absence of many of the motor cells. There was also a degeneration of the anterior root fibres, in many cases amounting to a complete absence of the nerve fibres. The crossed pyramidal tracts were markedly changed. Many of the fibres in the crossed pyramidal tracts seemed to be absent, and there appeared to be an increase in the con-

nective-supporting tissue. The direct or anterior pyramidal tracts were affected in a similar manner, although the pathological changes were not so marked in this region. The degeneration was not confined definitely to the pyramidal tracts, but extended out to the so-called mixed zone, and affected fibres which probably connect motor cells at different levels of the cord. For illustration of the described changes in cord, see Figs. 1, 2, 3.

The important point to which I wish to call your attention in this connection, is the seat of the pathological lesion in the cord; that is, that it was not con-

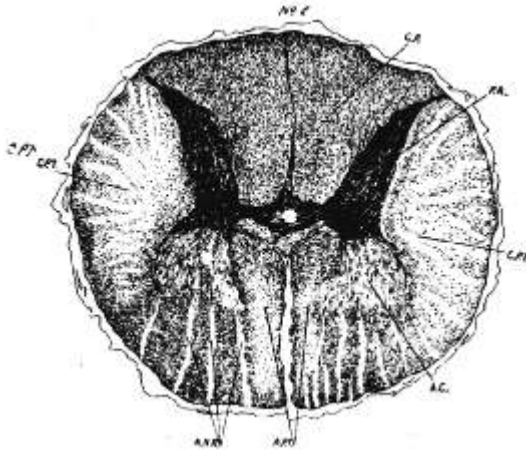


Section No. 1—Cervical Cord.

- C. P. T.* = Crossed pyramidal tracts, showing some degeneration of motor fibres.
- D. P. T.* = Direct pyramidal tracts, with slight changes.
- A. C.* = Anterior cornu, broken and having a granular appearance, with absence of some motor cells.
- A. N. R.* = Anterior nerve root fibres degenerated.
- P. C.* = Posterior columns, healthy.
- P. R.* = Posterior root, healthy.

fined to the lower segment of the motor path. Although there were no symptoms of any disease in the pyramidal tracts or the upper segments of the motor path, yet in this case there was a distinct and decided degeneration of the pyramidal tracts.

There is a class of cases coming under this head, the so-called amyotrophic lateral sclerosis of Charcot, in which we have weakness of the legs, with excessive myotatic irritability, while the muscles of the upper extremities are paralyzed and atrophied. The seat of the pathological changes in this class of cases is, as is well understood, in the anterior horn of gray matter of the cord, and the pyramidal tracts, the condition in the legs being explained by the changes in the pyramidal tracts, while the paralysis with atrophy of the upper extremities is due to changes in the anterior horn of



Section No. 2—Dorsal Cord.

C. P. T. = Crossed pyramidal tracts, markedly degenerated.

A. P. T. = Anterior pyramidal tracts.

A. N. R. = Showing absence of nerve root fibres.

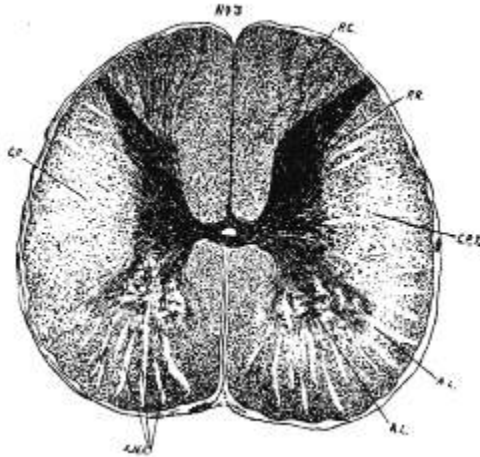
A. C. = Anterior cornu. Absence of many motor nerve cells, broken-down granular appearance.

P. C. = Posterior columns, having a healthy appearance.

P. R. = Posterior root, healthy.

gray matter and motor nerves of the cord. But the case under consideration does not come in this division. There were no symptoms during life indicative of any disease in the pyramidal tracts, or in the upper segment of the motor path; yet post-mortem examination showed these to have suffered from the morbid process.

The result of microscopical examination of the cord in this case, would then indicate that the seat of the lesion in the spinal cord in *atonic muscular atrophy* is not in the motor cells and motor roots only, but includes the pyramidal tracts as well. Gowers, of England, has recently shown that in these cases of *atonic muscular atrophy*, it is a very common occurrence to find the pyramidal tracts degenerated, not in the spinal cord alone, but this degeneration extending up through the pons and crus into the internal capsule of the brain, and



Section No. 3—Upper Lumbar Region.

- C. P. T.* = Crossed pyramidal tracts, degenerated.
A. C. = Anterior cornu, absence of motor cells and broken-down granular appearance of gray matter.
A. N. R. = Shows absence of anterior nerve root fibres.
P. R. = Posterior root, healthy.
P. C. = Posterior columns, healthy.

in some cases there was also a degeneration of the motor cells in the cortex of the brain.

Much might be said pro and con as to the seat of the primary lesion, whether in the pyramidal tracts or in the gray matter of the cord; of the possibility of the changes in the pyramidal tracts being secondary to those in the motor cells of the cord; or of the disease of

the lower segment of the motor path being secondary to the pathological changes in the pyramidal tracts.

We notice in this particular case that the changes in the pyramidal tracts and the gray matter of the cord seem to be of about equal intensity in the same section of the cord. In the lumbar region of the cord, where the motor cells were almost entirely destroyed and the anterior root fibres nearly absent, we find also a marked degeneration of the pyramidal tracts; while in the cervical region of the cord, which had to do with the muscles last paralyzed and wasted, the changes were not so extreme, and were of equal severity in the pyramidal tracts and motor cells of the cord. This would seem to indicate that the morbid process began in both pyramidal tracts and gray matter simultaneously, or at all events nearly so. The analysis of this case inclines us to the opinion that has before been expressed by Gowers, that *atonic muscular atrophy* is not an expression of disease and decay of the lower segment of the motor tract alone, but is, in many cases at least, an expression of the tendency to degeneration and decay of the whole motor path from the cortex of the brain to the muscles.

ON THE EFFECTS OF CHLOROFORM.

(British Med. Journal, Nov. 21, 1891.) L. E. Shaw, M.D., gives an account of some experiments made by Dr. Gaskell and himself which were in accordance with those of the Hyderabad Commission. They show that when chloroform is administered without interfering with perfectly regular respiration, complete insensibility can be produced without obvious weakening of the heart's beat. Inefficiency of the heart is brought about by the rapid inhalation of chloroform in too concentrated a form. The practical teachings are that it should be administered slowly and with plenty of air, and that great care should be taken not to push the chloroform when struggling or gasping respiration occurs.

A. F.

•